

## Case Report

# Thoraco-Omphalopagus Conjoined Twins at 29 Weeks Gestation – the Diagnostic and Management Challenges

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Conjoined twins are extremely rare, occurring in approximately 1% of monozygotic twins and having a birth incidence of approximately 0.2:10,000 births. Antenatal diagnosis of conjoined twins can be challenging and is normally made during antenatal surveillance with sonography in the first trimester in the UK. We present a case of thoraco-omphalopagus conjoined twins diagnosed at 29 weeks gestation after transfer of care from Nigeria. The case report discusses the diagnostic challenges of conjoined twins at this late gestation. We also explore the use of MRI imaging as an upcoming adjunct in the diagnosis of antenatal fetal problems. The counselling and antenatal planning resulted in this pregnancy being terminated and delivery by classical caesarean section eight days following the original diagnosis.

**ABBREVIATIONS**

MRI: Magnetic Resonance Imaging

**INTRODUCTION**

Conjoined twins are extremely rare, occurring in approximately 1% of monozygotic twins and having a birth incidence of approximately 0.2:10,000 births [1]. The prognosis is very unfavourable with most ending in miscarriage, stillbirth or early neonatal death.

The aetiology of conjoined twins is unknown with no established risk factors. The pathogenesis is also uncertain but if cell differentiation takes place after the 14<sup>th</sup> day of embryonic development, incomplete cleavage will result in the formation of conjoined twins. A system of classification established by St Hilaire in 1832 is still used today and is based on which structures are fused. The Greek suffix -pagus, meaning "what is fixed", differentiates the type.

Structural anomalies frequently associated with conjoined twins include polyhydramnios, cardiac malformations, common omphalocoels and neural tube defects [2].

Antenatal diagnosis of conjoined twins can be challenging and is normally made during antenatal surveillance with

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sonography. Signs indicating possible conjoined twins include a bifid appearance of the first trimester fetal pole, the presence of more than 3 umbilical cord vessels, failure of the fetuses to change position relative to each other over time and the persistence of the fetal heads at the same level and body plane [3]. The vast majority of reported cases of conjoined twins have been identified in the first or early second trimesters.

Because of the intrinsic limitation of ultrasound with regard to tissue contrast, MRI has been explored as a safe alternative and is fast becoming an important diagnostic tool used in the antenatal period for fetal assessment. In the first trimester MRI can be used for anatomical assessment and later in the pregnancy for prognosis, to plan delivery and evaluate for possible separation surgery.

**CASE PRESENTATION**

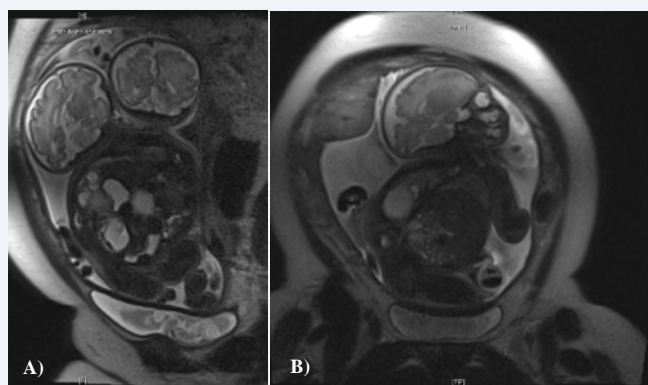
We present the case of a 34 year-old female from Nigeria who transferred her care to the Royal Bolton Hospital at 29 weeks gestation. A private scan in Nigeria at 20 weeks confirmed a mono chorionic twin pregnancy, with both twins lying transversely and their heads at opposite sides of the maternal abdomen. There was no comment as to whether this was a mono amniotic or di amniotic pregnancy and it is documented as 'nil abnormality seen'.

Her obstetric history included two previous vaginal deliveries of healthy babies at term and she had no past medical history of note. As she had recently transferred her care, she was referred for a growth/anomaly scan within the ultrasound department. On her first routine growth scan at the Royal Bolton Hospital, the ultrasonographer raised a suspicion of conjoined twins. This was confirmed by the local fetal medicine consultant and referred to the tertiary fetal medicine unit for further clarification.

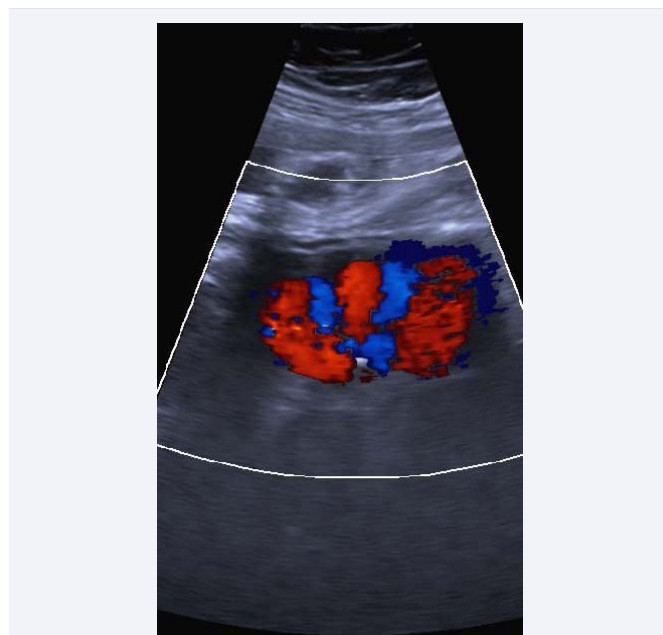
It was technically a very challenging scan and it was difficult to precisely identify the anatomy of the twins due to the advanced gestation. An MRI scan was subsequently undertaken to help clarify the anatomy. This confirmed thoraco-omphalopagus conjoined twins. The twins appeared to be sharing a single, poorly developed heart. There was a single cord insertion. Two kidneys were identified but only one appeared normally formed and deemed functional. It remained difficult to identify the



**Figure 1** Thoraco-omphalopagus conjoined twins at 29 weeks gestation following delivery.



**Figure 2** (A) – MRI of 29 week thoraco-omphalopagus twins (B) –MRI demonstrating tissue differentiation and allowing visualization of single cord insertion.



**Figure 3** Ultrasound of the single, poorly formed, shared fetal heart.

precise number of limbs but there appeared to be two legs, three arms and four hands. There were two distinct spinal cords.

The couple was counselled extensively about the poor prognosis associated with the ultrasound and MRI findings. The dangers associated with going into labour with conjoined twins in a transverse position were also discussed at length. They were offered termination of pregnancy and delivery by caesarean section or delivery by caesarean section and palliative care for the twins following delivery. They opted for termination of pregnancy and fetocide was undertaken using intra-umbilical potassium chloride. Unlike the umbilical cord, the heart was not easily accessible and this was the justification for opting for the intra-umbilical route.

The twins were delivered the following day, just 8 days after the original scan, by classical caesarean section, which was technically uncomplicated. The patient made an uneventful physical post-operative recovery. The couple wished for photographs to be taken and for their case to be used for teaching and training, but they did not want a post-mortem or any further investigation and the babies had a hospital burial.

## DISCUSSION

Conjoined twins are extremely rare, occurring in less than 1 in 100,000 pregnancies. They are monozygotic, mono chorionic and mono amniotic by definition, and are thought to result from incomplete embryonic division occurring between the 13<sup>th</sup> and 17<sup>th</sup> day after fertilization. The fetuses may technically be joined at any anatomical part and are categorized according to this. The most common form are thoracophagus (thorax) twins but can also be omphalopagus (abdomen), pygopagus (sacrum), ischiopagus (pelvis), craniopagus (cranium), cephalopagus (face) or rachipagus (back) according to the fusion site. Prognosis is difficult to determine and highly individualized, but depends mainly on the fusion site, how many and which organs are shared

and the presence of accompanying abnormalities. However, the overall outcomes are poor with up to 60% of conjoined twins demising antenatal and 35% are lost within the first 24 hours of life, mainly due to organ failure or during attempts to surgically separate them [2,4].

In the UK almost all conjoined twins are identified at the first trimester dating scan, with ultrasound findings described as 'fixed apposition of fetal bodies with fusion of skin lines'.

Although a catastrophic diagnosis at any gestation, early recognition allows for prompt referral to a fetal medicine unit, easier identification of anatomy with ultrasound, in depth counselling and planning and early termination of pregnancy if this is what the couple wishes. The aim is to minimize the psychological and physical effects to the mother.

Identification of conjoined twins in more advanced pregnancy, as in our case, brings a wealth of fresh problems. The precise anatomy can be difficult to accurately identify with ultrasound alone. This can be owed to reverberation artefacts in a more ossified fetus, diminished image quality in obese patients, oligohydramnios and engagement of the fetal head in late pregnancy [5]. This makes informed counselling, establishing prognosis and planning separative surgery more difficult. In our case, MRI scan was utilized to offer better tissue differentiation and allowed us to build a more detailed picture of the anatomical structure of the twins, which allowed us to offer a more informed discussion with regard to prognosis and antenatal planning. MRI is becoming a more utilized tool in the antenatal period. It is frequently used in the identification of fetal intracranial abnormalities due to its superior tissue differentiation properties allowing contrast between cerebral spinal fluid, brain tissue and

spinal cord. In such a rare condition such as conjoined twins, MRI proves to be a good complementary to ultrasound in complex anomalies.

Complex cases such as this require a very strong collaborative approach between the host hospital and tertiary level services. Counselling can often be difficult due the rarity of the condition but can be helped by complimentary diagnostics as previously discussed. If separative surgery was contemplated the involvement and counselling of neonatal surgical teams would be paramount.

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